

C A S E
R E P O R TSonographic Features of Nodular-type
Muscular Sarcoidosis

Hsin-Hua Chen¹, Tsu-Yi Hsieh¹, Der-Yuan Chen^{1,2,3*}, Howard Haw-Chang Lan^{3,4,5,6},
Chia-Wei Hsieh¹

Sarcoidosis is a granulomatous multisystem disorder that rarely involves muscle. Although characteristic ultrasound features of nodular-type muscular sarcoidosis have been reported, observations for such lesions at different disease stages are limited. We report ultrasonographic findings for nodular-type muscular sarcoidosis at 3 months and 1 year after the onset of disease in the left medial gastrocnemius muscle of a 35-year-old woman. A hypoechoic mass was initially detected in the involved muscle, and central echogenicity appeared later. It is, therefore, considered that ultrasonography can be a valuable tool in detecting and following up nodular-type muscular sarcoidosis lesions.

KEY WORDS — muscle nodule, muscular sarcoidosis, sonography

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Introduction

Sarcoidosis is a systemic granulomatous inflammatory disorder. Its clinical spectrum is protean, ranging from abnormal radiographic findings in an asymptomatic individual to severe multiorgan involvement. The incidence of symptomatic muscle sarcoidosis is 1.5–6%, while asymptomatic disease may occur in 5–80% of cases [1]. Three types of muscular sarcoidosis are described in the literature: acute myositic, atrophic, and nodular [2–6]. Although characteristic ultrasound (US) features of nodular-type muscular sarcoidosis have been reported [2,7,8], the US features with series follow-up have yet to be described. We report a case of nodular-type muscular sarcoidosis in the left medial gastrocnemius

muscle and describe the serial changes of US features prior to treatment.

Case Report

A 35-year-old woman noticed a firm, painless and immobile 4 cm mass in her left medial gastrocnemius muscle for 3 months before her first rheumatologic consultation. A 4–10 MHz linear-array US transducer (GE Logiq Book; Milwaukee, WI, USA) was used to examine the mass. A fusiform hypoechoic mass (3.8 cm in length) with hyperechoic striation was revealed in the left medial gastrocnemius muscle on the longitudinal scan (Fig. 1A). The mass had well-demarcated margins except at

¹Section of Allergy, Immunology and Rheumatology, Taichung Veterans General Hospital, Taichung, ²National Yang Ming University, Taipei, ³National Chung-Hsing University, ⁴Department of Radiology, Taichung Veterans General Hospital, ⁵Central Taiwan University of Science and Technology, and ⁶Hungkuang University, Taichung, Taiwan.

*Address correspondence to: Dr. Der-Yuan Chen, Section of Allergy, Immunology and Rheumatology, Taichung Veterans General Hospital, 160, Section 3, Chung-Kang Road, Taichung 407, Taiwan.
E-mail: dychen@vghtc.gov.tw

the two ends (Fig. 1). The rheumatologist was uncertain of the nature of the mass and suggested regular follow-up if the mass persisted or progressed. Seven months later, chest X-ray revealed bilateral hilar enlargement and interstitial infiltration over the bilateral lower lung fields. Computed tomography of the chest revealed multiple enlarged mediastinal and hilar lymph nodes. The patient was referred to the rheumatology section under the impression of sarcoidosis. At this time, another painless mass was noted in her right forearm. Whole-body gallium (Ga-67) scan disclosed increased isotope uptake over the mediastinum and bilateral lung hila (i.e. the positive lambda sign: a characteristic finding of sarcoidosis) and over the soft tissue of the left thigh, bilateral calves, right forearm,

and back. Follow-up US of the mass in the left calf using a 6–13 MHz linear array transducer (GE Logiq Series 500) revealed a striated mass that was hypoechoic at the periphery and hyperechoic at the centrum (Fig. 2). The mass was enlarged when compared with the previous sonographic image, and the middle portion occupied the entire layer of the gastrocnemius muscle. The margin between the mass and proximal adjacent muscle was clear; however, the border between the mass and distal adjacent muscle was blurred. Magnetic resonance imaging (MRI) (Siemens Sonata 1.5 Tesla; Erlangen, Germany) disclosed multiple lesions of variable size in the muscles of the left lower leg, with appearances of intermediate/high signal intensity on the T1-weighted image, high signal intensity on the

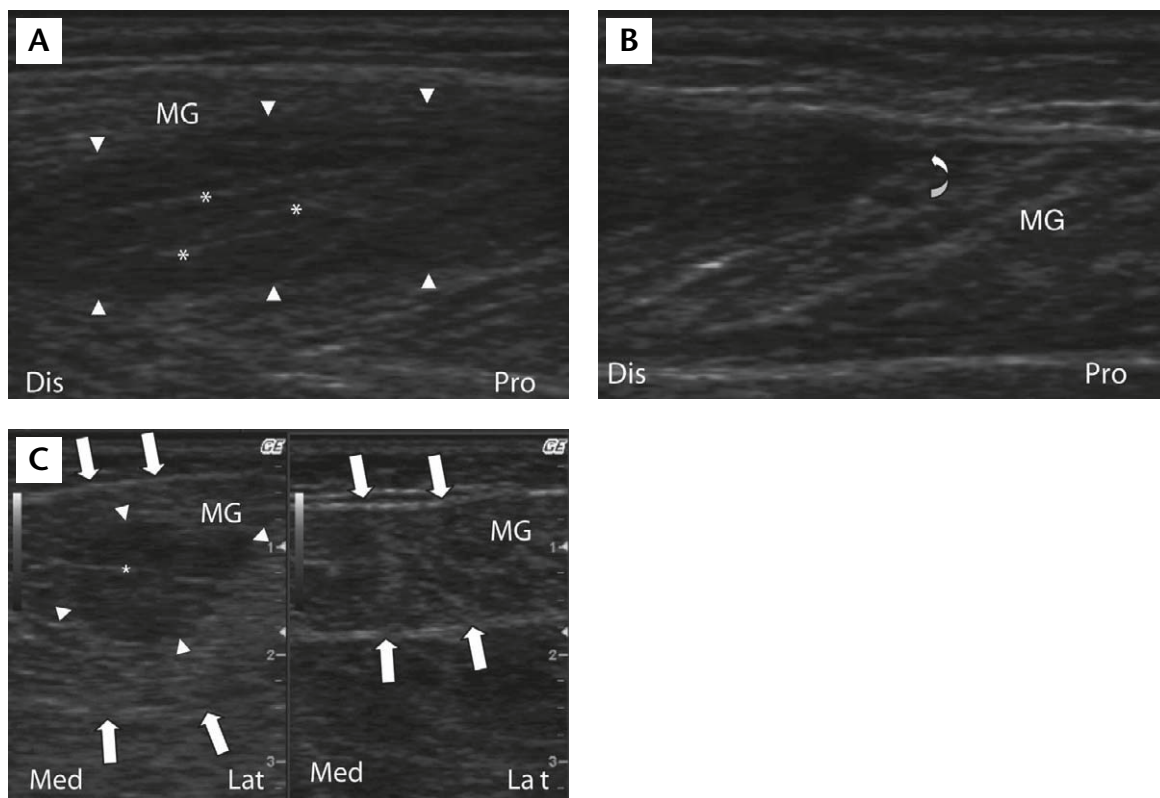


Fig. 1. Sonogram of the left calf. (A) Initial US performed 3 months after onset; longitudinal scan through the left medial gastrocnemius muscle. Note the hypoechoic mass lesion (between the arrowheads) in the central part of the left gastrocnemius muscle with some hyperechoic stripes (perimysium) at the centrum (*). The margin between the mass and adjacent muscle is clear (arrowheads). The margin at the two ends is not shown. (B) Upper part of the hypoechoic mass lesion. Note the ill-defined margin at the proximal end of the lesion (curved arrow). (C) Transverse scan of the mass through the left medial gastrocnemius muscle (left half), and right medial gastrocnemius muscle (right half) for comparison. Note the enlarged left medial gastrocnemius muscle (between the arrows). The margin between the mass lesion and adjacent muscle is clear (arrowheads). A hyperechoic stripe (*) is noted at the centrum. Dis = distal; Pro = proximal; Med = medial; Lat = lateral; MG = medial gastrocnemius muscle.

proton density image and T2-weighted image, and contrast enhancement on the T1-weighted image after intravenous administration of contrast agent (Gd-DTPA). On axial imaging, some lesions had a star-like area of low signal intensity with no contrast enhancement at the centrum (Fig. 3). The

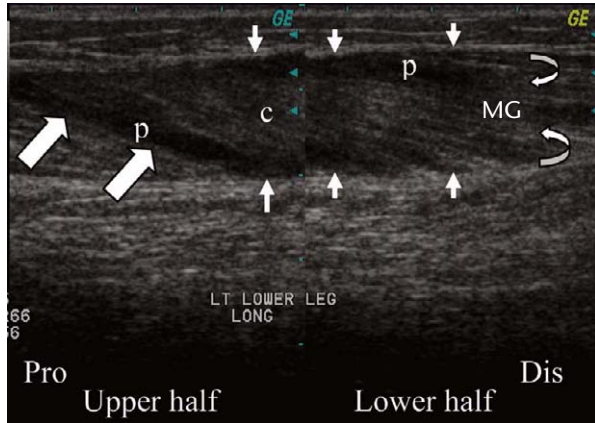


Fig. 2. Follow-up US performed 1 year after onset; longitudinal scan through the same muscle. Note the enlargement of the mass with involvement of the entire layer of the medial gastrocnemius muscle in some regions (between the arrowheads). The margin between the mass and adjacent medial gastrocnemius muscle is clear (arrows); however, the margin is ill-defined at its lower end (curved arrows). The mass reveals a characteristic appearance (arrows), i.e. hypoechoic at the peripheral area (p) and hyperechoic in the centrum (c) (perimysium). The centrum is more hyperechoic and larger than in the initial lesion. Pro = proximal; Dis = distal; MG = medial gastrocnemius muscle.

level of angiotensin-converting enzyme was elevated (46.2 IU/l/37°C; normal value, 8.3–21.4 IU/l/37°C), representing a diagnostic finding of sarcoidosis. An open biopsy of the mass in the left calf of the patient was performed. The histologic features revealed noncaseating granuloma, leading to a diagnosis of muscular sarcoidosis. The patient also displayed uveitis of the left eye that was proven by ophthalmic evaluation. After methylprednisolone pulse therapy (750 mg once daily for 3 days) followed by oral corticosteroid treatment, all muscle masses became impalpable and were negative to Ga-67 scan and MRI 3 months after treatment.

Discussion

Symptomatic muscle involvement of sarcoidosis has been reported to occur in 1.4% of known cases [9], but the relationship of the presence of intramuscular granulomas and clinical symptoms in muscular sarcoidosis is not yet known [10]. The nodular type is characterized by single or multiple masses that are usually noted in the extremities and that must be differentiated from soft-tissue neoplasms. The myopathic type involves muscles diffusely without forming a focal mass, and usually

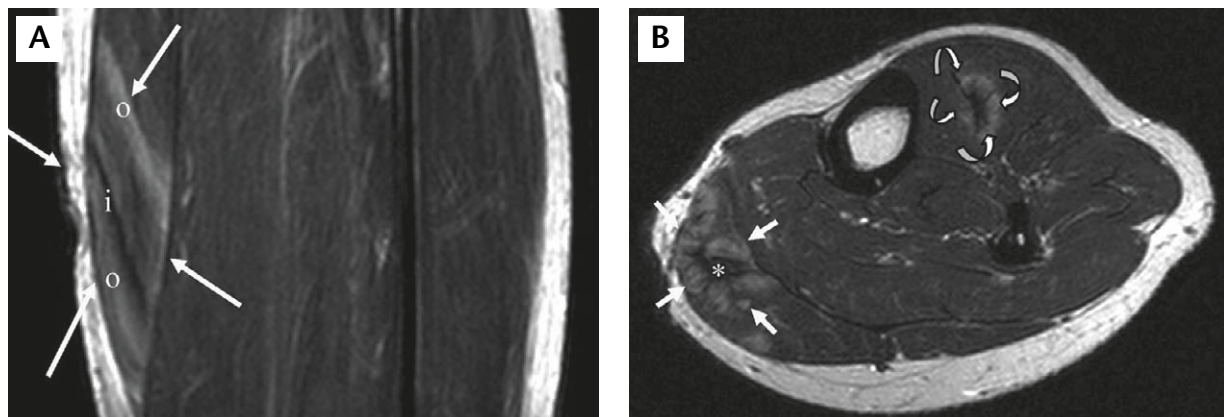


Fig. 3. Magnetic resonance imaging of the left leg; T1-weighted image with intravenous administration of contrast agent (Gd-DTPA). (A) Coronal scan through the calf muscles. Note the well-defined striated mass (arrows) with contrast-enhanced zones in the outer part (o) of the gastrocnemius muscle and zones of low signal intensity in the inner part (i). (B) Axial scan through the middle third of the lower leg. Note the corresponding mass lesion (short arrows) has a "dark star" low signal intensity at the centrum (*) and heterogeneous contrast-enhanced zones at the periphery. Note that another lesion (curved arrows) is located in the tibialis anterior muscle.

results in progressive weakness. The acute myositic type is the least common type, usually causing myalgia and tenderness [11].

Despite a 1-year history of glaucoma of the left eye thought to result from uveitis, our patient presented at the first rheumatologic consultation with a solitary painless palpable nodule in her left calf. The initial US of the mass showed a hypoechoic lesion with central hyperechoic striations, with a clear margin between the mass and adjacent muscle except at the two ends. The follow-up US 9 months later revealed a larger intramuscular mass. Previous studies reported the sonographic features of nodular-type muscular sarcoidosis [2,7,8]; however, the US findings of different stages of nodular muscular sarcoidosis prior to treatment have yet to be described. In the present study, the US features of the mass 1 year after its occurrence are similar to those in previous reports, showing inner stripes of hyperechogenicity and outer stripes of hypoechogenicity on the long axial view. According to a previous histopathologic study, the central areas of these masses are dense fibrosis (perimysium), while the peripheral areas are active inflammatory granulomas containing numerous epithelioid histiocytes [12]. On sonography, the hyperechoic part corresponds to the fibrous portion (perimysium) and the hypoechoic part to the active inflammatory granulomas. The lesion was fusiform along the muscle fibers and was located in the superficial part of the left calf, thus forming a palpable mass. At an earlier stage of the nodular type of muscular sarcoidosis, US might reveal only a few hyperechoic stripes at the centrum of a hypoechoic mass because of limited fibrosis, perhaps because there is little inflammatory granulomatous change in the early stage. As the disease progresses, the increasing inflammatory granulomatous change compresses the perimysium and results in thickened perimysium. These characteristic US features may be helpful in the diagnosis of nodular-type muscular sarcoidosis. These features can also be observed in nonspecific inflammatory granulomatous lesions or tuberculosis.

MRI of our case showed the characteristic appearance of a central star-shaped low-signal

area ("dark star" sign) in the muscle mass on all axial imaging, which corresponded to the US and pathologic findings (Fig. 3).

Ga-67 scintigraphy is useful in detecting sarcoidosis lesions that are not apparent on clinical examination and can direct physicians to perform biopsy over clinically occult sites of involvement for establishing the diagnosis. It is also a convenient method for following the course of intramuscular involvement in the management of sarcoidosis. In our case, Ga-67 scintigraphy revealed asymptomatic muscular involvement over the back and left thigh and was helpful in follow-up after corticosteroid treatment.

In conclusion, high-resolution US provides reasonably good tissue contrast in nodular-type muscular sarcoidosis, because the masses are usually located at the superficial portion of the muscles. It is more convenient and accessible and less expensive than MR and remains a useful tool in the initial evaluation of a palpable muscular nodule. Though the characteristic "central hyperechoic strip" appearance may not be prominent in the early stage of nodular-type muscular sarcoidosis, clinicians who are aware of its US features can perform early systemic survey for sarcoidosis and muscle biopsy to confirm the diagnosis.

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